

CASE REPORT

Exploring the uncommon : A case report of lymphangioma of a 8 year old child

Dr.Rubini Manavalan¹, Dr.Pournima Samayan Mohandass², Dr.G.Shanmugavadivel³, Joyson Moses⁴, Dr.A.Thirumagalanuraaga⁵, Dr.Hemcle Shalma G⁶, Dr.Karthikaivelan Jayaraman⁷, Dr.Ramya Raja⁸,

1. Post Graduate student, Department of Pediatric and Preventive Dentistry, Sri Venkateshwaraa Dental College, Puducherry 2. Assistant Professor, Department of Pediatric and Preventive Dentistry, Sri Venkateshwaraa Dental College, Puducherry 3. Professor & HOD, Department of Dentistry, Takshashila medical college hospital, Ongur, Tindivanam taluk, Villupuram 4. Professor and Head of Department, Department of Pediatric and Preventive Dentistry Sri Venkateshwaraa Dental College, Puducherry 5. Assistant Professor, Department of Pediatric and Preventive Dentistry, Sri Venkateshwaraa Dental College, Puducherry, 6. Reader, Department of Oral Medicine and Radiology, Sri Venkateshwaraa Dental College, Puducherry, 7. Assistant Professor, Department of Dentistry, Pondicherry Institute of Medical Science, Puducherry 8. Assistant Professor, Department of Pediatric and Preventive Dentistry, Sri Venkateshwaraa Dental College, Puducherry,

Corresponding author

Dr. Pournima Samayan Mohandass M.D.S.,

Assistant Professor, Department of Pediatric and Preventive Dentistry,

Sri Venkateshwaraa Dental College, Puducherry.

Exploring the uncommon: A case report of lymphangioma of an 8-year-old child

ABSTRACT

Lymphangiomas are rare, benign hamartomatous tumors of the lymphatic vessels that are most commonly found in the head and neck. There have been very few reports of lower lip incidence.

We present a rare instance of lower lip lymphangioma of an eight-year-old child who underwent surgical therapy but later relapsed at a 1 month follow up.

Keywords: Lymphangioma; Lower lip; Surgical excision; Follow-up

INTRODUCTION

A hamartoma is a benign, unifocal, or multifocal developmental abnormality composed of mature cells and tissues that are cytologically normal from the affected anatomical region. It has a disorganized architectural framework, with one aspect dominating the others. ⁽¹⁾

Hamartomatous forms known as lymphangiomas develop when aberrant lymphatic vessels fail to empty into adjacent lymphatics or veins, causing lymph to stagnate and the growth of massive, dilated lymphatic channels that resemble cysts. Lymphangiomas, which are regarded as non-neoplastic developmental anomalies rather than true tumors, are characterized by the aberrant proliferation of lymphatic vessels. ^(1,2)

Despite the fact that lymphangiomas are extremely uncommon in the oral cavity, they account for around 75% of all cases. The gingiva, palate, buccal mucosa, floor of mouth, and lips are hardly affected compared to the tongue, which is the most usually afflicted area inside the oral cavity. ^(3,4)

We present a unique instance of lymphangioma found in the lower lip of an 8-year-old child.

CASE DESCRIPTION

An 8-year-old girl presented to the Department of Pediatric and Preventive Dentistry with the chief complaint of a growth on her lower lip for the past 5 years. The patient's mother first noticed the growth when the child was 3 years old. It was initially small in size but gradually increased to its present size. The mother also mentioned that the child had the habit of biting the lesion. The lesion was painless, but the patient's main concern was the unsightly appearance of the lip. The patient had no previous medical and dental history.

On general examination, the patient was moderately built and nourished, with no signs of anemia, icterus, cyanosis, lymphadenopathy, or clubbing. On extraoral examination, there was no facial asymmetry, temporomandibular joint (TMJ) movements were within normal limits, and mouth opening was also normal.

A clinically confined, pebbly-surfaced, resembling cluster of vesicles with reddish hue, well-defined, asymptomatic, freely mobile mass on palpation and soft in consistency, measuring 2 x 1 cm, was discovered in the lower lip (Fig. 1). The lesion was deemed benign and provisionally diagnosed as a traumatic fibroma on the basis of the patient's mother's history.

The clinical presentation was taken into consideration for a differential diagnosis of lipoma and mucocele. With the parents' signed informed consent, an excisional biopsy (Fig. 2) with entire margin removal was performed chairside under local anesthesia, and the tissue sample was sent for histological examination.

The histopathological analysis (Fig. 3) revealed that the labial mucosa is lined by acanthotic stratified squamous epithelium. The subepithelium has intense mixed inflammation caused by neutrophils, lymphocytes, and plasma cells, as well as dilated lymphatic channels with fibrinous deposits, localized rupture, and granulation tissue formation.

One week later, the patient was reassessed (Fig. 4), revealing a small, smooth, and soft bump upon palpation. No treatment was necessary due to its minimal size. Follow-up evaluations were conducted at the third month and again at the sixth month (Fig. 5), where the lesion remained the same size as the previous visit, soft to the touch, and asymptomatic. No treatment was required as the lesion's size remained insignificant. The patient remains under follow-up.

DISCUSSION

Redenbacher was the first to define lymphangioma in 1828 ^(4,5) and Virchow was the first to characterize tongue lymphangiomas in 1854. ⁽⁴⁾ In children, these tumors make up 25% of all benign vascular tumors and 4% of all vascular tumors. A lack of evidence of gender or race discrimination has been found. ⁽⁵⁾ Lymphangiomas are rare, occurring in approximately 1 in 6,000 to 1 in 16,000 live births. In the pediatric population, these are accountable for about 9% of soft tissue tumors, maxillofacial tumors (11%), and benign tumors (6%). ⁽⁶⁾ Although it can also affect the axilla, inguinal, or vaginal areas, it mostly affects the head and neck region. Intraoral lymphangiomas, however, are uncommon. ⁽⁷⁾ Buccal mucosa is the next most often affected area in intraoral sites, after the anterior two-thirds of the tongue ⁽⁸⁾, however, in this case, it was discovered in the lower lip. Generally lymphangiomas manifest at birth (in 60-70% of cases) or by the time a child is two years old (90%). ⁽⁴⁾ Albeit they can also appear in adults as a result of trauma, inflammation, and lymphatic obstruction. ⁽⁵⁾ In contrast, our case report involves an 8-year-old girl with the condition with a 5-year prior history. Anjani Kumar et al. ⁽³⁾ reported a case similar to our case in a 13-year-old boy. Congenitally lymphangiomas can also be related to chromosomal abnormalities (such as Trisomy 21, Noonan syndrome and Turner syndrome), hemangiomas, fetal alcohol syndrome, hydrops fetalis, cardiac problems, and familial pterygium colli. ^(1,9) Although the precise cause of lymphangioma remains unclear, two significant ideas have been put up. As per the first notion, the head and neck region's lymphatic system is formed by endothelial out-pouching from the jugular sac, while lymphatic

channels evolve from five primitive sacs that originate from the venous system. In accordance with the second hypothesis, the lymphatic system is formed in part by mesenchymal clefts in the venous plexus reticulum that extend towards the center of the jugular sac. In cases where these enlarged primitive lymphatics are congenitally blocked or sequestered, lymphangiomas result.⁽¹⁰⁾ According to recent hypotheses, there is an increase of VEGF-C (vascular endothelial growth factor-C) expression and vascular endothelial growth factor R3.⁽¹¹⁾ Other etiological factors can include maternal viral infection, maternal substance abuse, and environmental factors.⁽¹²⁾

Clinically, oral lymphangioma appears as a gradually growing, painless lump located either superficially or within deep tissue.⁽⁷⁾ The surface appears pebbly, resembling a cluster of translucent vesicles, often likened to "frog eggs" or "tapioca pudding." Superficial lesions are typically described as papillary, which may either match the color of the surrounding mucosa or have a slightly reddish hue. Deeper lesions present as soft, ill-defined masses that appear as diffuse nodules or lumps, without notable changes in surface texture or colour.⁽¹⁴⁾ Histopathological features include lymphatic vessels that are markedly dilated, often diffusely infiltrating adjacent soft tissues, and exhibiting lymphoid aggregates within their walls. Lymphocytes and proteinaceous fluid fill the empty spaces in the thin endothelium lining. Secondary bleeding may develop within the lymphatic vessels. The lymphatic spaces contain neutrophils, macrophages, lymphocytes, red blood cells, and lymphatic fluid. There are a lot of inflammatory cells present in the loose fibrous tissue that makes up the surrounding connective tissue stroma.⁽¹⁵⁾ Similar histopathological features were observed in the present case with inflammatory infiltrate and granulation tissue secondary to a biting habit.

Three histological forms of lymphangiomas exist : ^(15,16)

- 1. Capillary Lymphangioma** consists of small capillary-sized vessels.
- 2. Cavernous Lymphangioma** consists larger dilated lymph channels.

3. Cystic Lymphangioma exhibits large macroscopic cystic spaces.

The differential diagnosis of lymphangioma includes several oral lesions, such as hemangioma, teratoma, dermoid cyst, thyroglossal duct cyst, amyloidosis, neurofibromatosis, heterotopy of gastric mucosal cyst, and granular cell tumor, all of which can result in macroglossia. Clinically, lipoma, salivary retention phenomenon, and hemangioma can be distinguished from lymphangioma. ^(4,9,11,16) Mucocele was ruled out in our case, as there were no mucin pools and an epithelial lining was present.

Diagnosis is primarily based on clinical presentation and histopathological examination, with imaging techniques such as ultrasound (USG) used to identify the cystic nature of the tumor. CT or MRI scans help determine the extent of the tumor and assist in surgical planning, while angiography may be required to exclude the likelihood of a hemangioma. ^(1,7,8,17)

Treatment for lymphangiomas depends on a variety of factors, including the type, size, involvement of anatomical structures, and extent of infiltration into surrounding tissues. Microcystic lesions are diffuse, do not respect tissue planes, and are challenging to remove, while macrocystic lesions are easily excised due to their confined nature ^(7,9,10,16).

There are various treatment modalities for lymphangiomas, including surgical excision, electrocautery, cryotherapy, radiation, intralesional steroids, sclerosing agents (such as OK432), embolization and ligation, Nd:YAG and carbon dioxide laser therapies, and radiofrequency tissue ablation. However, surgical excision is still the gold standard treatment. ^(1,3,4,7-10,12,13,15,16) In our case, since the diagnosis was initially made as a traumatic fibroma, we proceeded with a wide surgical excision under local anesthesia. Recurrence of lymphangioma is common because complete removal is challenging due to the lesion's non-encapsulated and "infiltrating" nature. ^(15,17,18)

CONCLUSION & CLINICAL SIGNIFANCE

Lymphangiomas of the lips are extremely rare. When treatment is necessary due to complications affecting the aesthetics or function of the lip, it is important to consider preserving lip integrity, function, and appearance. Although alternative therapy approaches have been proposed, surgical excision remains highly effective.



Figure 1: Lesion located in lower lip



Figure 2: Excisional biopsy

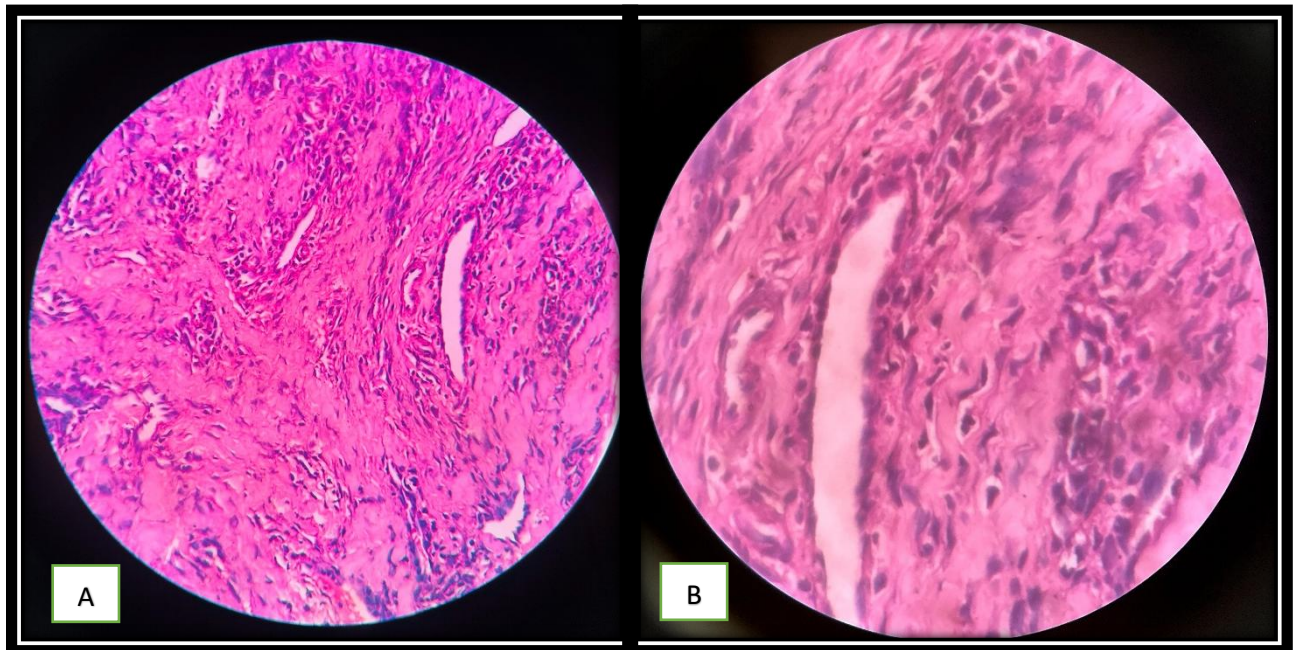


Figure 3: A – 40x magnification. B – 100x magnification. The labial mucosa is lined by acanthotic stratified squamous epithelium. The subepithelium has intense mixed inflammation caused by neutrophils, lymphocytes, and plasma cells, as well as dilated lymphatic channels with fibrinous deposits, localized rupture, and granulation tissue formation



Figure 4: Image showing 1 week review



Figure 5: Image showing 6 month review

REFERENCE

1. Hasan S, Ahmad SA, Kaur M, Panigrahi R, Panda S. Lymphangioma of the Lower Lip-A Diagnostic Dilemma: Report of a Rare Case with a Brief Literature Review. Case Rep Dent. 2022 Jun 2;2022:
2. Joshi PS, Hongal B, Sanadi A. Cystic lymphangioma: A differential diagnosis. J Oral Maxillofac Pathol. 2015 Sep-Dec;19(3):393-5.
3. Anjani Kumar JH, Madhumathi S, Anjan Kumar S, Pavan Kumar VV. Cavernous lymphangioma of the lower lip--a rare case report. Niger J Med. 2012 Oct-Dec;21(4):455-7.
4. Bhayya H, Pavani D, Avinash Tejasvi ML, Geetha P. Oral lymphangioma: A rare case report. Contemp Clin Dent. 2015 Oct-Dec;6(4):584-7.
5. Eren S, Cebi AT, Isler SC, Kasapoglu MB, Aksakalli N, Kasapoglu C. Cavernous lymphangioma of the tongue in an adult: a case report. J Istanbul Univ Fac Dent. 2017 Apr 3;51(2):49-53.
6. Matharu J, Patel V. Cryotherapy for the management of a lip lymphangioma – a case report and literature review. Oral Surg. 2019 Aug;12(3):230–7.
7. Dutta S, Ghosh DM, Sarangi S, Ray D, Ghosh R. Oral Lymphangioma: An Original Series of Three Cases with Review. Middle East Res J Dent. 2024 Jan 30;4(01):1–3.
8. Kurude AA, Phiske MM, Kolekar KK, Nayak CS. Lymphangiomas: Rare presentations in oral cavity and scrotum in pediatric age group. Indian J Dermatol Venereol Leprol. 2020 Mar-Apr;86(2):230.
9. V U, Sivasankari T, Jeelani S, Asokan GS, Parthiban J. Lymphangioma of the tongue - a case report and review of literature. J Clin Diagn Res. 2014 Sep;8(9):ZD12-4.

10. Akhtar K, Meraj F, Talat F, Haiyat S. Lymphangioma of the dorso-lateral tongue in an adult male: A rare case report. *IP J Diagn Pathol Oncol*. 2020 Dec 28;5(4):434–7.
11. Tayefeh A, Asadollahi A, Saatloo MV. A rare case report of oral lymphangioma in buccal mucosa. *Ann Clin Case Stud*. 2020;2(4):1035.
12. Chouchene F, Masmoudi F, Baaziz A, Maatouk F, Ghedira H. Oral manifestations and dental care management of a young patient with lymphangioma of the tongue: A case report. *Clin Case Rep*. 2021 Jul 23;9(7)
13. AlGhamdi A, AlBaqami M. Lymphangioma of the tongue in a pediatric patient. *J Pediatr Surg Case Rep*. 2021 Dec;75:102053.
14. Shetty DC, Urs AB, Rai HC, Ahuja N, Manchanda A. Case series on vascular malformation and their review with regard to terminology and categorization. *Contemp Clin Dent*. 2010 Oct;1(4):259-62.
15. Sunil S, Gopakumar D, Sreenivasan BS. Oral lymphangioma - Case reports and review of literature. *Contemp Clin Dent*. 2012 Jan;3(1):116-8.
16. Ugurlu F, Kaya A, Akdogan E, Olgac V, Sipahi A. Oral lymphangioma of the alveolar ridge – a report of a rare case. *Hamdan Med J*. 2016;9(3):247.
17. Khan S, Khan R, Ehtisham M, Nissar S, Gupta S. Lymphangioma of Lip: A Case Report. *Indian Journal of Contemporary Dentistry*, July-December 2020, Vol.8, No.2
18. de Carvalho FK, Pinheiro TN, da Silva RA, de Queiroz AM, da Silva LA, Nelson-Filho P. Lymphangioma of the Lower Lip Mimicking a Mucocele in Children. *J Dent Child (Chic)*. 2015 May-Aug;82(2):116-9.